Expected and unexpected cardiac problems during pregnancy

P.G. Pieper

Maternal heart disease complicates at least 1% of pregnancies and is one of the most important causes of maternal death. In developing countries rheumatic heart disease remains the major cause of maternal heart disease, while in developed countries maternal congenital heart disease has become more prevalent due to the improved survival of children with congenital heart disease. In addition, the prevalence of coronary artery disease is rising because of unhealthy lifestyle and the tendency to postpone motherhood until the third decade.

A report presenting data about maternal mortality in the UK from 2000 to 2002 showed that the majority of maternal deaths occurred in women with previously undiagnosed heart disease.[1] The haemodynamic burden of pregnancy can unmask previously asymptomatic heart disease. For example, it is not uncommon for women with rheumatic mitral stenosis to be diagnosed for the first time during pregnancy because the volume overload and increase in heart frequency cause symptoms that did not occur before pregnancy. The case reports of peripartum cardiomyopathy and coronary artery dissection in this issue illustrate that life-threatening heart disease can also develop as a new disease related to pregnancy.[2-4] In the reported cases the events occurred a few weeks before delivery or within days to weeks post-partum, but presentation several months after delivery is not uncommon. New cardiac disease can also coincide with pregnancy by chance as in the case report of a woman with third-degree AV block.[5] Both women with pre-existing yet undiagnosed heart disease and those with new (whether or not pregnancy-related) disease are likely to present primarily to obstetricians or primary caregivers and not to a cardiologist. A timely diagnosis, therefore, depends on adequate awareness on the part of their GP or obstetrician. The prevalence of heart disease in pregnant women and its potentially life-threatening consequences justify a careful cardiac history, family history and physical examination in all pregnant women. Recognising heart disease during pregnancy is challenging because the physiological changes during pregnancy can cause symptoms and signs mimicking cardiac disease. Examples are fatigue, shortness of breath, oedema and systolic ejection murmurs. Obstetric caregivers must be able to distinguish signs and symptoms attributable to normal pregnancy from cardiac pathology. Echocardiography and electrocardiography are the first-choice investigations that need to be performed when pathology is suspected. The threshold for referral to a cardiologist should be low.

Women with known heart disease before pregnancy need specialist pre-conception counselling. In fact, the risk of pregnancy must be assessed in teenagers with heart disease once they become sexually active. This is especially important when the risk for mother and/or foetus is significantly increased or when pregnancy is contraindicated. These teenagers need to be advised about safe and effective contraception. Combined hormonal contraceptives contain both oestrogen and progestagen and are attractive because of their reliable contraceptive effect. However, oestrogens enhance thrombogenicity which makes them unsuitable for women with increased thrombotic risk, even when they are on oral anticoagulation therapy. This applies to women with ischaemic heart disease, women with older types of mechanical valves and cyanotic women. Progestagen-only methods are suitable for these women. The mini-pill is, however, not recommended because of poor efficacy. Other progestagen-only methods such as depo-provera, cerazette and implanon are both effective and safe. The Mirena intrauterine system is safe when implanted with endocarditis prophylaxis, although concern has been raised for women who would poorly tolerate the vaso-vagal collapse that may accompany implantation (i.e. women with pulmonary vascular disease or Fontan circulation).[6]

P.G. Pieper
Department of Cardiology, University Medical Center Groningen, Groningen, the Netherlands

Correspondence to: P.G. Pieper
Department of Cardiology, Thorax Centre, University Medical Center Groningen, PB 30.001, 9700 RB Groningen, the Netherlands
E-mail: p.g.pieper@thorax.umcg.nl

When assessing pregnancy risk, existing risk scores must be implemented but also disease-specific risks must be taken into account (figure 1). Data to assist in providing an adequate risk assessment are available from several publications.[6-8] Contraindications for pregnancy are pulmonary hypertension, severely depressed systemic ventricular function (EF <30%), severe left heart obstruction, Marfan syndrome with ascending aorta >40 mm and peripartum cardiomyopathy with residual impairment of left ventricular function, because in these conditions maternal mortality is high.[6] Maternal risk is significantly increased in women with cyanotic heart disease, other complex congenital heart disease including Fontan circulation and systemic right ventricle, and mechanical prosthetic valves. When assessing maternal risk, it is important to realise that if several risk factors are present, these sometimes add up to an unacceptably high risk.[6,7]

A recent investigation among 116 women visiting a highly specialised congenital heart disease clinic demonstrated that 50% of women with a contraindication for pregnancy and over 30% of women with intermediate or high pregnancy risk did not recall being informed about the increased risk or contraindications.[9] This illustrates the importance of clear and repeated proactive counselling. Although pre-conception counselling should ideally start before transfer to adult cardiac services, the first visit to the adult cardiologist is particularly appropriate to inform the young woman about issues concerning pregnancy. Alternatively, a specialised nurse could provide this information. Young girls of 16 to 18 years may not be interested in receiving the complete information package, but there is a minimum amount of information that should be shared with them during this first visit. This should include information regarding the risk associated with pregnancy, whether pregnancy is contraindicated as well as information about the necessity to use safe and effective contraception. In addition, they should be advised to visit their cardiologist for full pre-pregnancy information before attempting to get pregnant. In due course, a full discussion about disease-specific maternal and foetal risks must take place. In congenital heart disease the inheritance risk should also be discussed and more often than not referral to a geneticist is appropriate.

In all but the heart diseases associated with the lowest risk (such as repaired ASD, VSD or patent duct, small ASD or VSD and mild pulmonary stenosis) the safety of pregnancy for both mother and foetus will improve when pregnancy is carefully planned. A full clinical investigation is indicated before pregnancy. This includes physical examination, oxygen saturation, electrocardiogram, echocardiogram and exercise testing. When systemic ventricular function cannot be accurately assessed echocardiographically, MRI should be performed. Twenty-four hour electrocardiography may be useful in women with a high risk of ventricular arrhythmias. Pregnancy risk assessments can be optimised using the results of these investigations. In women with aortic dilatation or severe valvular disease pre-pregnancy surgery must be considered, but the high risk associated with pregnancy in women with a mechanical valve prosthesis must be weighed against the risks associated with native valve disease.[10] The case report of two women with long-QT syndrome and pregnancy-related torsades de pointes illustrates that in such women pre-pregnancy ICD implantation is a sensible policy.[11] The impact on foetal well-being of any medication the woman uses must be appreciated. Although medication doses should be kept as low as possible and some medication should be avoided, the mother-to-be needs to understand that discontinuing necessary medication will not only negatively affect her but also her baby's health. For example, refusal to use anticoagulation for prevention of mechanical valve thrombosis may result in 200% mortality (mother and baby). ACE inhibitors, angiotensin II antagonists and statins are not compatible with pregnancy and need to be stopped or replaced. Beta-blocking agents (pre-

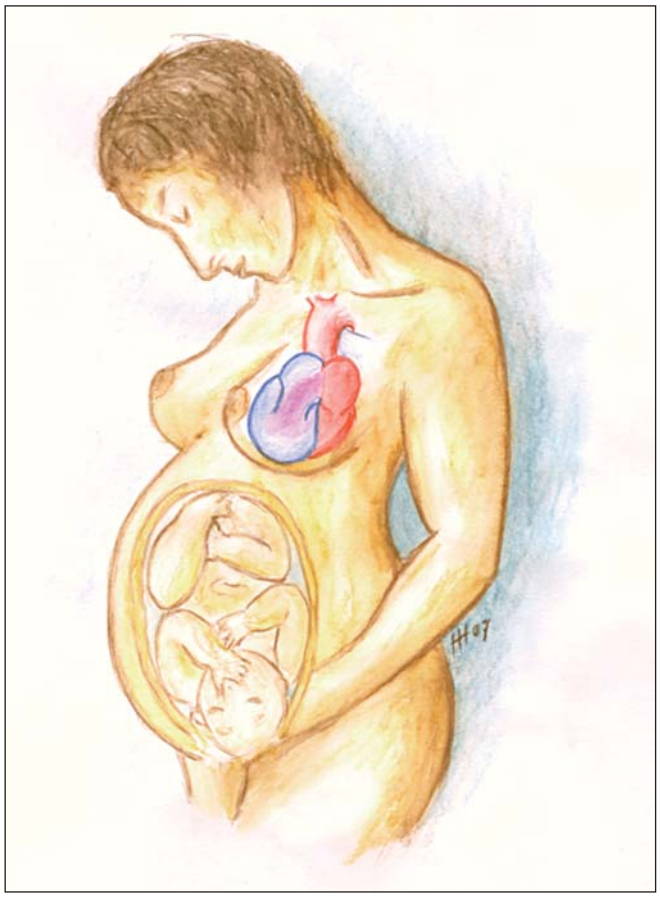

Figure 1. A pregnant woman with large uncorrected atrial-septal defect (ASD). Although pregnancy is generally well tolerated in women with ASD, the risk of thromboembolic complications and arrhythmias is increased (courtesy J.P.M. Hamer).[8]

ferably metoprolol), diuretics (except spironolactone) and digoxin are relatively safe. The risks and advantages of different anticoagulation regimens must be discussed with women who need anticoagulation and agreement must be reached between the physician and the woman concerning the preferable regimen.[10] Before pregnancy a decision should be made regarding the preferred level of care during pregnancy: predominantly in a local hospital for the most simple lesions, in a specialist cardiology and obstetric clinic for severe, complex and rare heart disease, and shared care between these two options in lesions of intermediate complexity.

Substandard care was a contributing factor in 40% of cardiac maternal deaths in the UK maternal mortality report.[1] Substandard care included poor communication between responsible specialists and lack of clearly defined management strategies. In order to avoid such problems a multidisciplinary team should be available to attend to women with cardiac disease once pregnancy is established. The minimum team requirement should be a cardiologist, an obstetrician and an anaesthesist with experience in cardiac disease. Paediatric expertise will be necessary as well, and the involvement of specialised nurses can be valuable. It is advisable to schedule a cardiological and obstetric consultation early in pregnancy, and subsequent visits at least each trimester but more often in complex or severe disease. Reports from each consultation must be immediately provided. These reports should not only cover a description of the current clinical situation and therapy, but also an assessment of problems that can be expected later in pregnancy such as arrhythmias and heart failure and their management. A plan for the management of delivery must be in place at the end of the second trimester.

Education of cardiologists and obstetricians as well as anaesthesists is necessary to optimise care for pregnant women with cardiac disease and should be part of their standard curriculum. Two excellent and up-to-date textbooks are available for practitioners who want to expand their knowledge about this increasingly important subject.[12,13] ■

References

1 Why Mothers Die 2000-2002. Report on Confidential Enquiries into Maternal Deaths in the United Kingdom. Royal College of Obstetricians and Gynaecologists; London: 2004.
2 Bosch MGE, van der Voort PHJ, Bams JL, Santema JG. A serious complication in the puerperium: peripartum cardiomyopathy. *Neth Heart J* 2008;**16**:415-8.
3 Oosterom L, de Jonge N, Kirkels JH, Klöpping C, Lahpor JR. Left ventricular assist device as a bridge to recovery in a young woman admitted with peripartum cardiomyopathy. *Neth Heart J* 2008;**16**:426-8.
4 Van den Branden BJL, Bruggeling WAJ, Corbeij HMA, Dunselman PHJM. Spontaneous coronary artery dissection in the postpartum period. *Neth Heart J* 2008; **16**:412-4.
5 Tietge W, Daniëls M. A case of an acquired high-degree AV block in a pregnant woman. *Neth Heart J* 2008;**16**:419-21.
6 Thorne S, MacGregor A, Nelson-Piercy C. Risks of contraception and pregnancy in heart disease. *Heart* 2006;**92**:1520-5.
7 Siu SC, Sermer M, Colman JM, Alvarez AN, Mercier LA, Morton BG, et al. Cardiac Disease in Pregnancy (CARPREG) Investigators. Prospective multicenter study of pregnancy outcomes in women with heart disease. *Circulation* 2001;**104**:515-21.
8 Drenthen W, Pieper PG, Roos-Hesselink JW, van Lottum WA, Voors AA, Mulder BJ, et al. Outcome of pregnancy in women with congenital heart disease: a literature review. *J Am Coll Card* 2007; **49**:2303-11.
9 Kovacs AH, Harrison JL, Colman JL, Sermer M, Siu SC, Silversides CK. Pregnancy and contraception in congenital heart disease: what women are not told. *J Am Coll Card* 2008;**52**: 577-8.
10 Pieper PG, Balci A, van Dijk AP. Pregnancy in women with prosthetic heart valves. *Neth Heart J* 2008;**16**:406-11.
11 Meregalli PG, Westendorp ICD, Tan HL, Elsman P, Kok WEM, Wilde AAM. Pregnancy and the risk of torsades de pointes in congenital long-QT syndrome. *Neth Heart J* 2008;**16**:422-5.
12 Oakley C, Warnes CA, editors. Heart Disease in Pregnancy. Second edition. Blackwell Publishing 2007.
13 Steer PJ, Gatzoulis MA, Baker Ph. Heart Disease and Pregnancy. RCOG Press, London, 2007.